Leprosy

Dr. Nashwan Mansoor

Leprosy

- Leprosy (Hansen's disease) is a chronic granulomatous disease affecting skin and nerves.
- Caused by *Mycobacterium leprae.*
- *A* slow-growing mycobacterium that cannot be cultured in vitro.
- The clinical manifestations are determined by the degree of the patient's cell-mediated immunity towards *M. leprae*.
- High levels of CMI with elimination of leprosy bacilli produces tuberculoid leprosy.
- Absent CMI results in lepromatous leprosy.
- Complications arise due to nerve damage, immunological reactions and bacillary infiltration.

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- People with leprosy are frequently stigmatized and using the word ‘leper’ is inappropriate.
- Untreated lepromatous patients discharge bacilli from the nose.
- Infection occurs through the nose, followed by hematogenous spread to skin and nerve.
- The incubation period is 2–5 years for tuberculoid cases and 8–12 years for lepromatous cases.
- Leprosy incidence peaks at 10–14 years.
- More common in males and in household contacts of leprosy cases.

Leprosy

❖ Pathogenesis :-

- M. leprae has tropism for Schwann cells and skin macrophages.
- In tuberculoid leprosy, effective CMI controls bacillary multiplication and organized epithelioid granulomata form.
- In lepromatous leprosy, there is abundant bacillary multiplication, e.g. in Schwann cells and perineurium.
- Borderline tuberculoid, in patients with moderate CMI.
- Borderline lepromatous, in patients with little cellular response.

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❖ Pathogenesis :-

- Immunological reactions evolve as the immune response develops and the bacillary antigenic stimulus varies, particularly in borderline patients.

- Delayed hypersensitivity reactions produce type 1 (reversal) reactions, while immune complexes contribute to type 2 (erythema nodosum leprosum) reactions.

- HIV/leprosy co-infected patients have typical lepromatous and tuberculoid leprosy skin lesions and typical leprosy histology and granuloma formation.

- Even with low circulating CD4 counts, tuberculoid leprosy may be observed and there is not an obvious shift to lepromatous leprosy.

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❖ Clinical features :-

➢ **The cardinal features of leprosy includes the following ;-**

- ❑ **Skin lesions, typically anesthetic at tuberculoid end of spectrum.**
- ❑ **Thickened peripheral nerves.**
- ❑ **Acid-fast bacilli on skin smears or biopsy.**

➢ **Types of leprosy are:-**

- ❑ **Lepromatous leprosy.**
- ❑ **Tuberculoid leprosy.**

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❖ Clinical features :-

➢ Skin.

- ❑ The most common skin lesions are macules or plaques.
- ❑ Tuberculoid patients have few, hypopigmented lesions.
- ❑ Lepromatous leprosy, papules, nodules or diffuse infiltration of the skin.
- ❑ The earliest lesions are ill defined; gradually, the skin becomes infiltrated and thickened.
- ❑ Facial skin thickening leads to the characteristic leonine facies.

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❖ Clinical features :-

➢ Anesthesia.

- ❑ The small dermal sensory and autonomic nerve fibers are damaged, causing localized sensory loss and loss of sweating.
- ❑ Can occur in the distribution of a damaged large peripheral nerve.
- ❑ A ‘glove and stocking’ sensory neuropathy is also common in lepromatous leprosy.

➢ Nerve damage.

- ❑ Peripheral nerve trunks are affected at ‘sites of predilection’ .
- ❑ Damage to peripheral nerve trunks produces characteristic signs with regional sensory loss and muscle dysfunction.

- ❑ All these nerves should be examined for enlargement and tenderness, and tested for motor and sensory function.
- ❑ The CNS is not affected.

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❖ Clinical features :-

➢ Eye involvement.

- ❑ Blindness is a devastating complication for a patient with anesthetic hands and feet.
- ❑ Eyelid closure is impaired when the facial nerve is affected.
- ❑ Damage to the trigeminal nerve causes anesthesia of the cornea and conjunctiva.
- ❑ The cornea is then susceptible to trauma and ulceration.

➢ Other features.

- ❑ Many organs can be affected.
- ❑ Nasal collapse occurs secondary to bacillary destruction of the nasal cartilage and bone.
- ❑ Diffuse infiltration of the testes causes testicular atrophy and the acute orchitis that occurs with type 2 reactions.
- ❑ This results in azoospermia and hypogonadism.

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❖ Leprosy reactions :-

❑ Type 1 (reversal) reactions.

- ✓ **Occur in 30% of borderline patients.**
- ✓ **Are delayed hypersensitivity reactions.**
- ✓ **Skin lesions become erythematous.**
- ✓ **Peripheral nerves become tender and painful.**
- ✓ **Sudden loss of nerve function.**
- ✓ **May occur spontaneously, after starting treatment and also after completion of multidrug therapy (MDT).**

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❖ Leprosy reactions :-

❑ Type 2 (erythema nodosum leprosum, ENL) reactions.

- ✓ **Due to immune complex deposition.**
- ✓ **Occur in BL and LL patients who produce antibodies and have a high antigen load.**
- ✓ **Manifest with malaise, fever and crops of small pink nodules on the face and limbs.**
- ✓ **Iritis and episcleritis are common.**
- ✓ **Other signs are acute neurites, lymphadénites, orchites, bone pain, dactylites, arthritis and proteinuria.**
- ✓ **May continue intermittently for several years.**

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❖ Borderline cases :-

- Skin lesions are more numerous than in tuberculoid cases.
- more severe nerve damage and a risk of type 1 reactions.

- In borderline leprosy (BB) cases, skin lesions are numerous and vary in size, shape and distribution; annular lesions are characteristic and nerve damage is variable.

- In borderline lepromatous (BL) cases, there are widespread small macules in the skin and widespread nerve involvement; both type 1 and type 2 reactions occur.

- Pure neural leprosy occurs principally in India and accounts for 10% of patients.
- Asymmetrical involvement of peripheral nerve trunks and no visible skin lesions.

- On nerve biopsy, all types of leprosy have been found.

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❖ Investigations :-

- **The diagnosis is clinical, made by finding a cardinal sign of leprosy.**
- **Supported by detecting acid-fast bacilli in slit-skin smears or typical histology in a skin biopsy.**
- **Smears are useful for confirming the diagnosis and monitoring response to treatment.**
- **Neither serology nor PCR is sensitive or specific enough for diagnosis.**

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❖ Management :-

➢ Principles of leprosy treatment treatment

- ❑ **Stop the infection with chemotherapy.**
- ❑ **Treat reactions.**
- ❑ **Educate the patient about leprosy.**
- ❑ **Prevent disability.**
- ❑ **Support the patient socially and psychologically.**

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❖ Management :-

➢ All leprosy patients require MDT with an approved first-line regimen.

➢ Rifampicin :-

✓ A potent bactericidal for M. leprae.

✓ Should always be given in combination with other antileprotics, since a single-step mutation can confer resistance.

➢ Dapsone:-

✓ Bacteriostatic.

✓ Commonly causes mild hemolysis and rarely anemia.

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❖ Management :-

➢ Clofazimine :-

- ✓ **A fat-soluble crystalline dye, weakly bactericidal for M. leprae.**

- ✓ **Skin discoloration (red to purple–black) and ichthyosis are troublesome side-effects, particularly on pale skins.**

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❖ Management :-

- The agents are now established second-line drugs;-

✓ Fluoroquinolones (pefloxacin and ofloxacin) are new bactericidal drugs against *M. leprae*.

✓ Minocycline and clarithromycin may also be used.

- Minocycline causes a grey pigmentation of skin lesions.
- The single-dose treatment is less effective than the conventional 6-month treatment for paucibacillary leprosy,
- Chloroquine can also be used.

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❖ Management :-

➢ **Lepra reactions :-** are treated with Prednisolone for Type1 reaction, and prednisolone or thalidomide for Type2 reaction.

➢ **Patient education :-**

- ❑ The patients should be informed that, after 3 days of chemotherapy, they are not infectious and can lead a normal social life.
- ❑ It should emphasize that gross deformities are not inevitable.
- ❑ Patients with anesthetic hands or feet need to avoid and treat burns or other minor injuries.
- ❑ Good footwear is important.

➢ **Physiotherapy :-** helps maintain range of movement of affected muscles and neighboring joints.

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❖ Prognosis :-

- Untreated, Tuberculoid leprosy has a good prognosis; it may self-heal and peripheral nerve damage is limited.
- Untreated, Lepromatous leprosy (LL) is a progressive condition with high morbidity.
- Borderline patients are at risk of developing type 1 reactions, which may result in devastating nerve damage.
- After treatment, the majority of patients,, do well, with resolution of skin lesions, especially those who have no nerve damage at the time of diagnosis.

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❖ Prevention and control :-

- **The previous strategy of centralized leprosy control campaigns has been superseded by integrated programmed, with primary health-care workers in many countries now responsible for case detection and provision of MDT.**

- **Is not yet clear how successful this will be, especially in the time-consuming area of disability prevention.**

- **BCG vaccination has been shown to give good but variable protection against leprosy; adding killed M. leprae to BCG does not enhance protection.**

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